

Amendments to the Claims:

This listing of claims replaces all prior versions and listings of claims in the application:

Listing of Claims:

1. **(Currently Amended)** A method of identifying a candidate therapeutic compound for treating facioscapulohumeral muscular dystrophy (FSHD), the method comprising[[,]] :

- (a) providing a D4Z4 binding element comprising DNA comprising one or more 4q35 D4Z4 repeats;
- (b) contacting the D4Z4 binding element with a test compound;
- (c) determining whether the test compound interacts with the D4Z4 binding element; and
- (d) ~~selecting the test compound as a candidate therapeutic compound for treating FSHD if the test compound interacts with the D4Z4 binding element;~~
- (e) providing a cell comprising a D4Z4 binding element operably linked to a FSHD region gene 1 (FRG1);
- (f) contacting the cell with the selected test compound;
- (g) determining the level of FRG1 expression in the cell in the presence of the test compound;
- (h) comparing the level of FRG1 expression in the cell in the presence of the test compound to a reference level of FRG1, and
- (i) selecting the test compound as a candidate therapeutic compound for treating FSHD if the level of FRG1 expression in the cell in the presence of the test compound is reduced as compared to the reference level of FRG1.

2. (Currently amended) The method of claim 1, wherein the cell comprises an endogenous D4Z4 binding element is in a cell that expresses and operably linked FSHD region gene 1 (FRG1).

3.-4. (Cancelled)

5. (Currently amended) The method of claim [[2]] 1, wherein the cell is a muscle cell.

6. (Currently amended) The method of claim [[5]] 1, wherein the cell is from a subject that has FSHD.

7. (Original) The method of claim 1, wherein the interaction is the binding of the test compound to the D4Z4 binding element.

8. - 9. (Cancelled)

10. (Withdrawn, Currently amended) ~~The method of claim 1, further comprising,~~
A method of identifying a candidate therapeutic compound for treating facioscapulohumeral muscular dystrophy (FSHD), the method comprising:

- (a) providing a D4Z4 binding element comprising DNA comprising one or more 4q35 D4Z4 repeats;
- (b) contacting the D4Z4 binding element with a test compound;
- (c) determining whether the test compound interacts with the D4Z4 binding element; and
- (d) selecting the test compound if the test compound interacts with the D4Z4 binding element;

- [[(a)] (e) providing a D4Z4 binding element (DBE) comprising DNA comprising one or more 4q35 D4Z4 repeats and a D4Z4 recognition complex (DRC) under conditions such that the DBE and the DRC can bind to each other;
- [[(b)] (f) contacting the D4Z4 binding element and DRC or a DRC component with [[(a)] the selected test compound identified by the method of claim 1; and
- [[(c)] (g) determining whether the test compound affects the binding of the D4Z4 binding element to the DRC or DRC component,

wherein an increase in binding between the D4Z4 binding element and the DRC or DRC component in the presence of the test compound indicates that the test compound is a candidate compound for treating FSHD.

11. (Withdrawn) The method of claim 10, wherein the DRC component is YinYang 1 (YY1), High Mobility Group Box 2 (HMGB2), or nucleolin.

12. - 20. (Cancelled)

21. (Currently amended) The method of claim 1, wherein the D4Z4 binding element is in a reporter construct comprising A method of identifying a candidate therapeutic compound for treating facioscapulohumeral muscular dystrophy (FSHD), the method comprising [(a)] :

- (a) providing a cell expressing a D4Z4 binding element reporter construct comprising DNA comprising one or more 4q35 D4Z4 repeats, operably linked to a promoter and a reporter gene;
- (b) contacting the cell with a test compound;
- (c) determining a level of expressing of the reporter construct in the presence of the test compound;

- (d) comparing the level of expression of the reporter construct in the presence of the test compound to a reference representing a level of expression in the absence of the test compound; and
- (e) selecting the test compound as a candidate therapeutic compound for treating FSHD if the level of expression of the reporter construct in the presence of the test compound is reduced as compared to the reference.

22. (Cancelled)

23. (Previously presented) The method of claim 21, wherein the reporter construct comprises one, two or six minimal D4Z4 binding elements.